Superior sinus venosus atrial septal defect with variant and anomalous right pulmonary venous return

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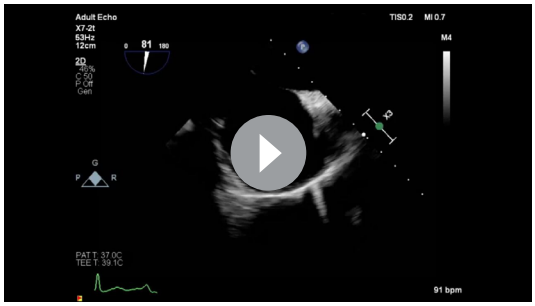
DESCRIPTION

A previously healthy woman in her 30s was referred to our hospital due to progressive dyspnoea and bilateral leg oedema for 2 months. Physical examinations showed a leftward and downward shift of cardiac apex, wide and fixed split S2, and a grade III/VI systolic ejection murmur at the left upper parasternal border area. Mild pitting oedema of both legs was also noted. Other examinations including chest auscultation were unremarkable. A 12-lead electrocardiography appeared normal. Chest radiograph in posteroanterior view and upright position showed an increased cardiothoracic ratio and pulmonary blood flow as well as the prominent size of the main pulmonary trunk and right descending pulmonary artery. Transthoracic echocardiography (TTE) revealed the right atrial (RA) and right ventricular dilatation with preserved systolic function (figure 1A,B). The pulmonary artery pressure was slightly elevated with an estimated right ventricular systolic pressure of 54 mm Hg. There was no evidence of significant valvular abnormality or intracardiac shunt on TTE. Since the likelihood of intracardiac shunt was high, subsequent transoesophageal echocardiography was



Video 1 Transoesophageal echocardiography in mid-oesophageal, bicaval view showed a 1.8 cm echogenic drop-out of an interatrial wall at the superior vena cava and right atrium junction.

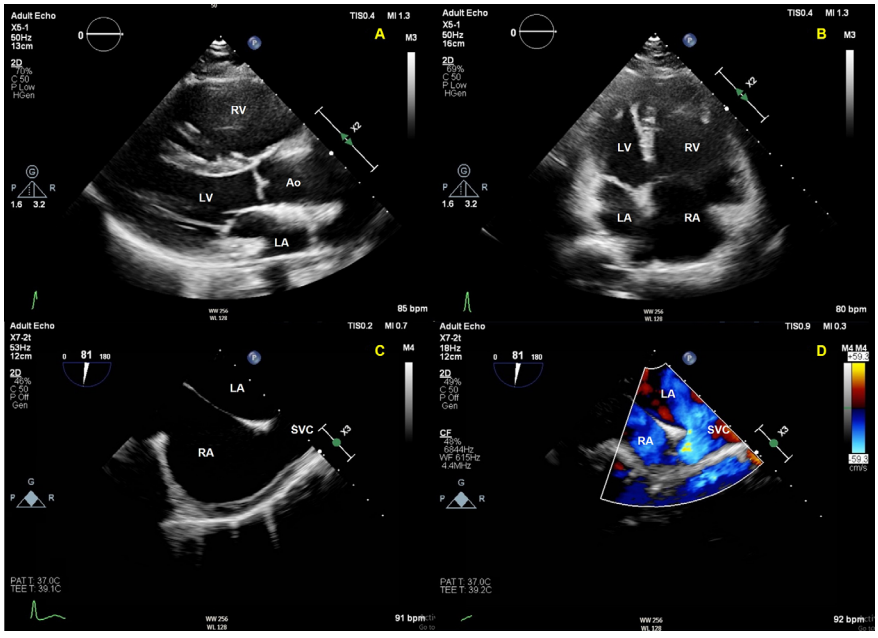


Figure 1 Transthoracic echocardiography in parasternal long axis view (A) and apical four-chamber view (B) showed RA and RV dilatation. Transoesophageal echocardiography in mid-oesophageal, bicaval view (C) and (D) showed a 1.8 cm echogenic drop-out of an interatrial wall at the SVC-RA junction with blood flowing from LA to RA, indicating superior sinus venosus atrial septal defect with left-to-right shunt. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

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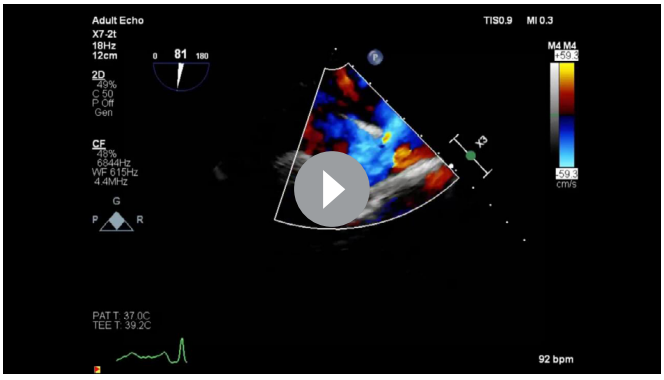


Video 2 Transoesophageal echocardiography with colour flow study in mid-oesophageal, bicaval view showed that the blood flowed from the left atrium to the right atrium.

performed and demonstrated a 1.8 cm echogenic drop-out of an interatrial wall at the superior vena cava (SVC)-RA junction, indicating superior sinus venosus atrial septal defect (SVASD) (figure 1C and video 1). The colour flow study showed that the blood flowed from the left atrium (LA) to RA, a left-to-right shunt (figure 1D and video 2). All four pulmonary veins (PV) drained to the LA. Cardiac MR (CMR) imaging also demonstrated SVASD and depicted coexisting partial anomalous pulmonary venous returns (PAPVR) from the right upper and one branch of right middle PV to SVC (draining 4.4 cm and 1.7 cm above the

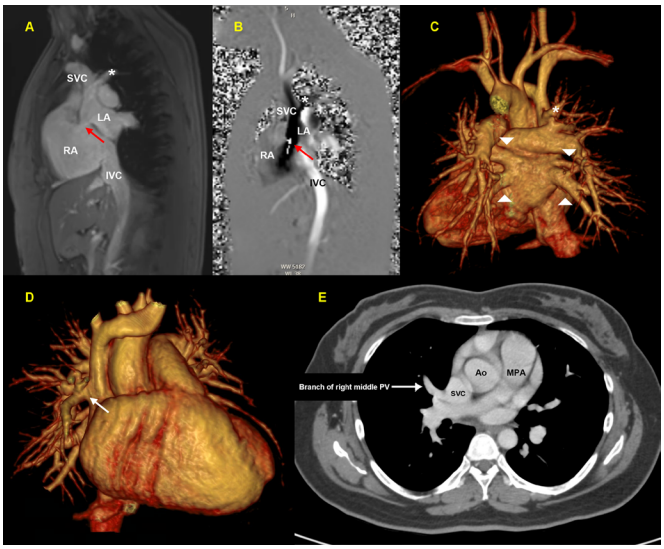
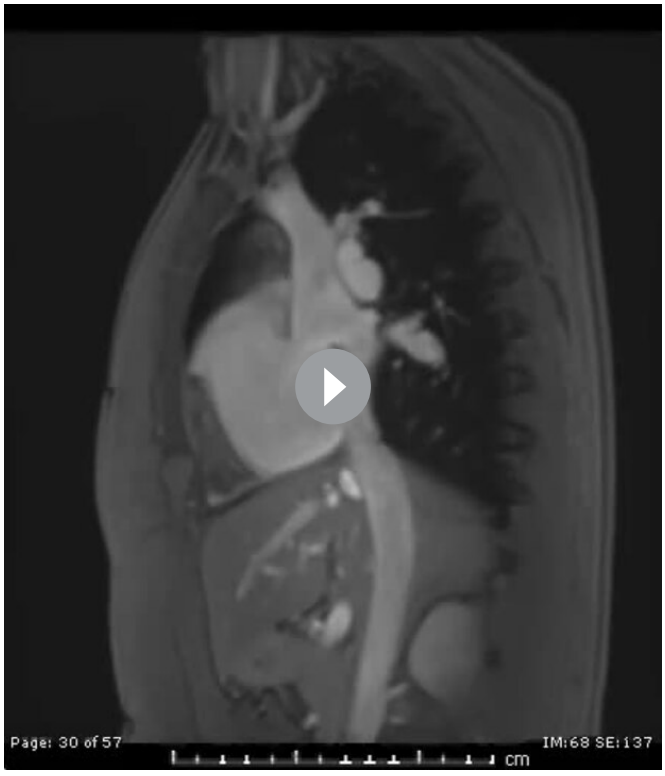


Figure 2 CMR with steady-state free precession cine (A) and phase-contrast imaging (B) in bicaval view showed superior sinus venosus atrial septal defect (red arrow) and anomalous right upper PV return to SVC (asterisk). CMR with volume rendering technique in posterior view with the removal of descending thoracic aorta (C) showed 4 PVs draining into LA (arrowheads) and an anomalous right upper PV return to SVC (asterisk). CMR with volume rendering technique in anterior view (D) showed an anomalous branch of right middle PV return to SVC (white arrow). CT pulmonary angiography in axial view (E) showed an anomalous branch of right middle PV return to SVC (white arrow). Ao, aorta; CMR, cardiac MR; IVC, inferior vena cava; LA, left atrium; MPA, main pulmonary artery; RA, right atrium; PV, pulmonary vein; SVC, superior vena cava.



Video 3 Cardiac MR imaging with steady-state free precession cine in bicaval view showed superior sinus venosus atrial septal defect and anomalous pulmonary venous return to superior vena cava.

lower end of SVASD, respectively) while another branch of right middle, right lower and both left PV drained normally to LA (figure 2A–C and videos 3 and 4). SVC over-rode the interatrial wall into LA; however, the majority of SVC and anomalous PV flow was drained into RA as shown in figure 2B and video 4. The estimated pulmonary-systemic perfusion ratio (Qp:Qs) was 3.2. A CT pulmonary angiography from the referring hospital was reviewed and clearly showed PAPVR (figure 2D,E). Given the patient's symptoms and evidence of significant haemodynamic consequences from multimodality imaging, the correction was indicated either by surgery with the Warden procedure or transcatheter covered stent placement. Due to our centre of expertise, surgical correction with the Warden procedure was scheduled after meticulously patient informed consent.

SVASD is a rare type of atrial septal defect (ASD) representing 5%–10% of all ASD cases.[1] Most SVASD cases (80%–100%) often coexist with PAPVR which typically arises from the right lung (80%).[2–4] Given its accessibility, accuracy and relatively non-invasiveness, echocardiography, either a transthoracic or transoesophageal approach, is the first-line investigation for ASD. However, echocardiographic evaluation of sinus venosus ASD with PAPVR can be challenging due to the limitation of pulmonary venous drainage assessment. Most of the population have 4 PV (right upper, right lower, left upper and left lower) draining into LA via four independent ostia, although, 30% of cases have PV variations in the number and draining ostia, particularly right PV (figure 3).[5 6] Therefore, the presence of four PV draining into LA cannot exclude PAPVR as demonstrated by our case. Additional investigations, for example, cardiac CT and CMR, can

Video 4 Cardiac MR imaging with phase-contrast in bicaval view showed superior sinus venosus atrial septal defect and anomalous pulmonary venous return to superior vena cava.

be helpful for completing pulmonary venous drainage system assessment. Moreover, CMR has some advantages over other imaging modalities in terms of cardiac volume and shunt quantification. Surgical correction, the Warden procedure, is recommended in patients with impaired functional capacity or enlarged right cardiac chamber size from a significant left-to-right shunt (Qp:Qs≥1.5) as the current standard of care.

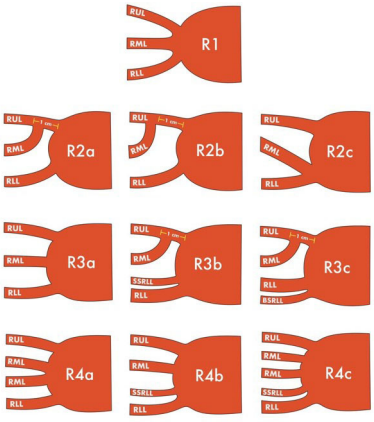


Figure 3 Demonstrated variation of the right pulmonary venous drainage patterns (adapted, with permission, from figure 1 in Marom *et al*. Variations in pulmonary venous drainage to the left atrium: implications for radiofrequency ablation. Marom EM,[6] p824, RSNA 2004). BSRLL, basilar segment right lower lobe; RLL, right lower lobe; RML, right middle lobe; RUL, right upper lobe; SSRLL, superior segment right lower lobe.

Furthermore, transcatheter correction with covered stent increasingly performs as an alternative treatment in patients with suitable anatomy (eg, the anomalous PV directly connects with the LA and no other large PV anomalously drains into high SVC).[7] Although the anatomy of defects, in this case, seems to be able to correct with both techniques, the patient was scheduled to undergo the Warden procedure due to the experience of our centre. This case highlights the importance of multimodality cardiovascular imaging for evaluating sinus venosus ASD with PAPVR.

Patient's perspective

It was shocking to know that there are many holes in my heart. Fortunately, all of these holes can be closed.

Learning points

- Given that more than 80% of sinus venosus atrial septal defect (ASD) coexists with partial anomalous pulmonary venous return (PAPVR), a complete pulmonary venous drainage system should be carefully evaluated.
- There are many pulmonary venous system variations in the number and draining ostia; therefore, four pulmonary venous draining into LA cannot exclude PAPVR.
- In addition to echocardiography, multimodality cardiovascular imaging, for example, cardiac CT and cardiac MRI, should be considered when assessing patients with sinus venosus ASD and suspected PAPVR.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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